BURR (C. W.)

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I PROPOSE to study with you to-day a disease, or rather a group of diseases, characterized by progressive muscular wasting and paralysis, caused by degeneration of the muscles themselves, and not by a lesion of the nervous system. The affection, therefore, does not properly belong, in a strictly scientific classification, among nervous diseases, but since its symptoms are all nervous and its differential diagnosis is from diseases of the cord and nerve-trunks, we have full justification for its study.

At first glance the boy, D. F. (Fig. 1.), looks remarkably robust. The calves are large, the bony frame well developed, the facial expression intelligent, and the complexion ruddy. But, as you see, he cannot stand, can, indeed, scarcely move the legs at all, and while in the arms power of movement in all directions is preserved, yet the movements are awkward, and cannot be made against even slight resistance. The neck-muscles, on the other hand, are fairly strong, and those of the face perfect, so that he can smile, and frown, and wink without difficulty. On examining the muscles more closely we find that while some are enlarged, are apparently hypertrophied, others are small, are distinctly atrophied. The calves, loins, and infra-spinati are large, hard, and firm. The thighs are soft and flabby, but not small, while the shoulder-muscles, the lower part of the great pectorals and the muscles of the upper arms are dis-

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tinctly softer and smaller than normal. The forearms are wasted but little, if any, and the intrinsic muscles of the hands not at all. Many of the muscles, those enlarged most and those most atrophied, give no elec-

Fig. 1.

Pseudo-hypertropic muscular paralysis.

tric reaction to either the faradic or the galvanic current, and in those which do react the reaction is reduced to both currents. Reaction of degeneration is

nowhere present. Your first thought, no doubt, was that the tremendously enlarged calves and loins indicated great strength, but you can now see that these are the weakest parts of the body.

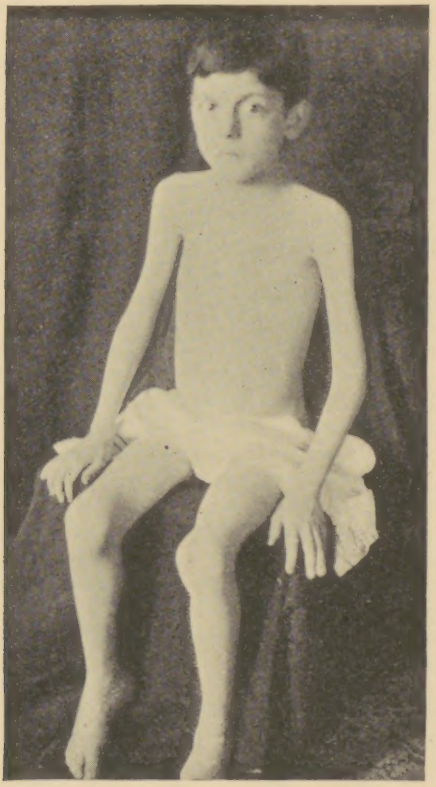
The mechanical reflex of the muscles, the contraction obtained by percussion of the muscle itself, is everywhere abolished, even in the great pectorals, in which in health it is ordinarily most marked and most readily obtained. Not only is there no contraction of the muscle in its entirety, but also the local contraction at the seat of percussion, the "humping" of a few fibers, is absent. The knee-jerk, and indeed all the tendon-jerks, are absent. Nowhere do we observe any fibrillary twitching. Sensation is normal to touch, pain, and temperature. The special nerves are good. The patient has complete control of the bladder and rectum. There are no trophic changes in skin, bones, or joints. There is slight talipes equinus on both sides. Subjectively the boy complains of nothing but muscular weakness. He eats well, digests well, sleeps well, and suffers no pain.

I have here a second boy affected in the same way, but to a less degree, so that he is still able to walk, and I wish you to watch his gait and method of getting up from the ground and lying down, for it is characteristic. You notice as he stands his feet are placed widely apart, his shoulders thrown back, and his abdomen forward, producing a large anterior curvature of the lower spine. His gait is waddling, duck-like, the body being twisted and thrown to one or the other side, corresponding to the foot which is upon the ground. Most characteristic of all, watch him as he rises. He rolls over on his stomach, plants his toes firmly against the floor, gets "on all fours," and then putting his hands with a jerk, one after the other, on knees and thighs, pushes the trunk up. As has been well said by Gowers, he climbs up his own legs. On lying down he goes through precisely the reverse procedure, as you see.

The third boy, L. X. (Fig. 2), presents a striking con-

trast to the others, and is an example of a much rarer condition. He looks like a living skeleton. The skin is

Fig. 2.



Simple idiopathic muscular atrophy.

too big for the body, and hangs loosely from it. The wasting is well-nigh universal, affecting not only arms,

legs and trunk, but also (and note this particularly) the face, as is shown by the projecting eyeballs, the slightly protuberant lower lip, the inability to pout, the curiously dull expression called "the myopathic face," due to the absence of some of the facial lines. Except for the absence of enlarged muscles and the involvement of the face, his symptoms are similar to those of the other boys, and his condition the same.

Since now we have learned all we can from examination of the patients, for I know nothing of their previous lives, let us consider for a few moments the causation and mode of progress of the disease. You probably wonder why I have presented the third case as an example of the disease affecting the other two. Great as the contrast is, the dissimilarity is to a large degree superficial, and in all the underlying cause is the same. Clinically, however, we separate them, calling the one pseudo-hypertrophic muscular paralysis, the other simple idiopathic muscular atrophy. The latter has been subdivided into several types according to the distribution of the wasting. Thus there is the juvenile type of Erb, in which the muscles of the shoulders, upper arms, and thigh, and the gluteal muscles are affected; and the facio-scapulo-humeral type, in which the face is attacked first. Other subdivisions have been made, but we may without loss disregard them. No hard-and-fast line can be drawn between any of the types, since mixed cases are met with.

The disease is developmental, due to a fundamental defect in the tissues that form muscle, but what the defect is and its cause we do not know. The affection is not congenital in the sense of showing symptoms at birth, but it is congenital in origin. It occurs far more frequently in family groups than in isolated cases. In the hypertrophic form there is never, or almost never, direct inheritance from parent to child, but transmission is through the healthy female line; that is to say, women who are themselves not affected may yet, if one may say so, possess the potentiality of the disease. In the simple

form there is greater possibility of direct inheritance, and females are far more frequently affected. Pseudo-muscular hypertrophy begins in late infancy or early childhood, idiopathic atrophy a little later, and sometimes, though rarely, in adult life. The course of events in pseudo-hypertrophy is as follows: The child, who has before walked, will begin to stumble and may complain a little of pain. He tires easily, and says his legs are weak. Now, at the first, this is all, and no diagnosis can be made, though by the exclusion of other diseases we may be able to more than shrewdly guess what will be forthcoming. Later, the calves begin to be enlarged and firm, and muscular weakness increases slowly but surely, until a child, apparently a young giant, is in truth a helpless weakling. All muscles do not enlarge. The pseudo-hypertrophy is usually in the calves, the extensors of the legs, the glutei, the loins, deltoids and infra-spinati. In rare cases the tongue is affected. Certain muscles, as for example the lower part of the great pectorals and the latissimus dorsi, atrophy without previous enlargement. As time passes the enlarged muscles also atrophy, and contortions and contractions appear, the legs being flexed upon the thighs, the thighs upon the abdomen. In the simple form the symptoms may begin, as already stated, in the face, shoulders, or legs.

To sum up, we may say that idiopathic muscular atrophy is a chronic, inherited, developmental disease characterized by progressive wasting and paralysis, with or without muscular hypertrophy, gradually diminishing tendon-reflexes and muscle-reflexes, gradually diminishing electric reaction, without trophic changes, with normal sensation, and with complete control of bladder and rectum.

Using this as a working definition, let us see how we can differentiate this affection from wasting due to other causes. What are the other causes? First, and for our purpose least important, is wasting due to cerebral disease. For a long time it was denied that this existed,

but of late years several cases have been reported in which tumors of the motor area, without disease of the anterior cordal gray matter, were accompanied by wasting in the palsied parts, and I do not mean the merely slight wasting due to disuse, but distinct atrophy. We are absolutely ignorant of the cause of this condition, but its distribution, hemiplegic or monoplegic, and the preceding palsy, make it impossible to be mistaken for idiopathic atrophy. Wasting from bulbar disease also needs only to be mentioned. Its accompanying symptoms differentiate it. Most important and most difficult is the diagnosis from atrophy due to spinal-cord disease, the so-called myelopathies. Any disease involving degeneration of the motor cells in the anterior gray matter will show wasting in the muscles with which those cells are connected. Hence it follows that wasting is found in many cordal diseases and with varying accompanying symptoms, according to what other parts of the cord are involved. All in which increased tendon- and muscle-reflexo-jerks and fibrillary twitching occur can be at once excluded, since, as we have seen in idiopathic atrophy, the so-called myopathies, the reflexes gradually diminish until they become extinct. The affection that most nearly simulates a myopathy is chronic anterior poliomyelitis, or progressive spinal muscular atrophy. But this is a disease almost exclusively of adult life. The wasting begins in the thenar and hypothenar eminences, or deltoid muscle, and while the tendon-jerks are abolished unless the lateral tracts be affected, in which case they are exaggerated, the muscle-jerks are increased and fibrillary twitching is common. There may be one of several changes in the electric reactions, including reaction of degeneration. Acute anterior poliomyelitis may, if first seen some time after the attack, deceive at first sight, but the acute onset, the reaction of degeneration, and the distribution of the palsy, in one leg or arm, or two or even three extremities, suffices to differentiate it. In chronic multiple neuritis the method of onset and progress, the

distribution, the frequent sensory symptoms, the possible ataxia, the palsy more intense than the wasting, are all points in differential diagnosis.

In myopathies there is ordinarily an inheritance; there are likely to be several cases in a generation; the onset is early in life; trophic changes never occur, while in myelopathies inheritance plays no part; the disease almost invariably begins later in life; trophic changes are not infrequent, and hypertrophy never occurs.

Senator has described a form of subacute or chronic myositis—an inflammation, you understand, not a degeneration—in which there is paralysis and wasting, but the the intense pain from pressure on the muscles and the course of the disease differentiate it.

Lastly, to complicate matters, there is a type of wasting likely to occur in family groups, and beginning early in life in the extensors of the big toe, appearing later in the common extensors and peroneal group, and thence extending. Reaction of degeneration and fibrillary twitching may or may not be present, and hypertrophy never occurs. It is the so-called peroneal type of Charcot and Tooth. It is probable that the situation of the lesion is not constant, but it may be in the anterior motor cells of the cord, the nerve-trunks or the muscles, since on no other grounds can we explain the varying electric reactions and reflexes.

The prognosis as to recovery in pseudo-muscular hypertrophy is absolutely bad. Death is almost inevitable within a few years after the patient becomes bedridden. In simple atrophy the course of the disease is longer, and there may be arrest, and the patient may reach old age.

Treatment is absolutely without curative influence. Electricity and massage will, however, do much to keep patients in the best possible condition, and gymnastics, when possible, may stimulate the development of what little muscular tissue is present. Drugs so far have proved useless.